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ART. I.—*Notice of a Case of Cyanosis, or the Blue Disease, connected with mutual adhesion of the semilunar Valves of the Pulmonary Artery.* By DAVID CRAIGIE, M. D., &c., Senior Physician to the Royal Infirmary. (Read at the Fourth Evening Meeting of the Royal College of Physicians.)

SINCE the time at which William Hunter* and Dr Pulteney† recorded their cases of direct communication between the right and left sides of the heart, and solicited the attention of physiologists to the consequent effect of that communication, in preventing the proper coloration of the blood in the lungs, and the other changes required by the healthy condition of that fluid, many different instances of different degrees of *Cyanosis* have been published, all tending to throw more or less light upon the lesion in a physiological and pathological point of view.

The case published at Leyden by Sandifort, in 1777, which had been seen by Gaubius in the year 1769, and by Hahn in 1776, attracted much attention, and suggested to that anatomist various interesting and important observations, which have been more or less fully repeated by subsequent authors. This case does not, however, appear to have been known to English physicians till long after its publication, as no mention is made of it till after the lapse of several years.‡

For the first systematic views on this lesion we are indebted to Corvisart, who, in his classical work on the Diseases of the Heart, has collected chiefly from a dissertation published by M. Cailliot of Strasbourg, several cases, previously little known, in illustration

* Medical Observations and Inquiries, Vol. vi. Art. xxvii. 1783.

† Transactions of the College of Physicians, Vol. iii. Art. xxi. 1785.

‡ Ed. Sandifort, Observationes Anatomico-Pathologicae, Lugduni Bat. 1777. Cap. I. de rarissimo Cordis vitio.

of the pathological varieties which the lesion assumes.* From these facts it resulted that the symptoms known, not very accurately, under the name of *Cyanosis* and *Morbus Cæruleus* might depend not only on, 1st, the open or patent state of the *foramen ovale*, but on perforation of the *septum cordis*, congenital or acquired; 2d, on the aorta arising in such a manner that its orifice corresponds to a congenital aperture in the septum; 3d, on the aorta arising from the right and left ventricle at once, as in the case recorded by Sandifort and Dr Nevin, and the 47th case of M. Gintrac, the case of M. Olivry; and the case recorded by Dr George Gregory; 4th, on the pulmonary artery arising from the left ventricle, while the aorta arose from the right, as in the case recorded by Baillie; or 5th, on the aorta and pulmonary artery rising from the left ventricle, as in the case recorded by M. Marchale; or 6th, on the circumstance of there being only one auricle and one ventricle, the latter giving rise to one trunk, which afterwards divides into the aorta and pulmonary artery.

An account, at once more complete, and at the same time digested with greater precision, was soon after given in 1816, by Kreyzig, Professor of Medicine at Dresden, who observed the occasional contraction or narrowing of the pulmonary artery, which, though in some cases seen, had not been often noticed. He is the first who appears to have attached due importance to the arctation or closure of the pulmonary artery.† Other monographical treatises on this lesion are, a dissertation published by Charles Frederic Haase at Leipsic in 1813,‡ and another published at Gottingen in 1816, by John Charles Hein.§ About the same time, viz. in 1814, M. Gintrac published on these cases a good dissertation, which he, in 1824, expanded into a complete monograph, embracing all the cases known up to that time.|| He has devoted particular attention to the state of the pulmonary artery, which was more or less contracted or narrowed in sixteen of fifty-three cases, and in five more obliterated. The latter lesion, it may be believed, is rare; because it is difficult to conceive how life can be continued, even with great arctation, much less obliteration of the pulmonary artery. In point of fact, of the five cases presenting what M. Gintrac calls obliteration, one was in an infant who lived only seven days, the case communicated by Mr Hodgson to Dr Farre; another took place in a child who died in the fifth month, whose

* Essai sur les Maladies et les lésions Organiques du Cœur, &c., 4trieme Classe, Art. iii., §. II., p. 303. Paris, 1818.

† Die Krankheiten des Herzens systematisch bearbeitet und durch eigene Beobachtungen erläutert von D. Friedrich Ludwig Kreyzig. Berlin, 1816. Zweiter Theil Zweite Abtheilung, S. 825.

‡ Dissertatio Med. Inaug. Medica de Morbo coeruleo. Lipsiae, 1813.

§ De istis Cordis deformationibus quæ Sanguinem venosum cum arterioso misceri permittunt. Gottingae, 1816.

|| Observations et Recherches sur la Cyanose, ou Maladie Bleu; par E. Gintrac, D. M., &c. Paris, 1824. 8vo.

case is recorded by Mr Howship ; and in the third, which is one of those given by Dr Farre, the infant lived only five weeks.

Meanwhile, the subject had attracted the attention of M. Louis, who, in 1823, published in the Archives Generales de Medecine his views on the lesion, in a memoir on the communication of the right and left cavities of the heart. This author takes no notice of the dissertations either of Haase, of Hein, or of his countryman Gintrac ; nor does he in the republication of his memoir in 1826 mention the existence of the monograph of the last author, though that had appeared two years previously,—in 1824. M. Louis distinguishes the different forms of the lesion into five varieties ; 1st, communication of the auricles by means of the *foramen ovale* left patulous ; 2d, communication of the ventricles by means of perforation of the septum ; 3d, communication between the right and left cavities of the heart by means of the *ductus arteriosus* and the *foramen Botalli*, or by means of perforation of the septum of the ventricles ; 4th, communication between the auricles and the ventricles by means of apertures in their partitions ; and 5th, communication between the right and left cavities of the heart by means of the *foramen Botalli*, and by the aorta arising from the two ventricles.

In his subsequent general view of the pathological relations of the preternatural communication he attaches considerable importance to the different forms of contraction of the pulmonary artery. The origin of these he traces to different periods. Some, as those which are formed by a perforated diaphragm, he thinks must be congenital ; others, however, he regards as of more recent formation ; and he finally infers that it coincides in more than one-half of the cases with a very marked arctation of the pulmonary artery, which may be considered as congenital.*

From studying the more extensive set of cases afterwards collected by M. Gintrac, and comparing the histories of those contained in the writings of British physicians, with the appearances presented by the remarkable case recorded in the present paper, I have been led to draw conclusions regarding the pathological relations of the several lesions usually found in the case of communication of the right and left side of the heart, much more decided regarding the fact of contraction of the pulmonary artery, than are given, either by Kreysig, by Gintrac, or by Louis. I have been led to think that some degree or form of contraction, or arctation of the pulmonary is not only the first, but the most essential lesion ; and that the open state of the foramen of Botallus, or the perforation of the ventricle, are to be regarded as dependent on this contraction, and, in a great measure, determined by this contraction having previously taken place to a degree, so great, as to

* Memoires ou Recherches Anatomico-Pathologiques. Par P. Ch. A. Louis, D.M., &c. Paris, 1826. P. 301.

render the free and natural circulation through the pulmonary artery impracticable.

There are three forms of lesion to which the pulmonary artery is liable; the first is a very unusually contracted formation of its cylinder; the second is complete obstruction of its interior; and the third is more or less occlusion of its interior by coalescence and mutual adhesion of its valves. From the cases hitherto recorded the two first lesions appear to be congenital; but it is not easy to say whether the third is congenital, or depends on some morbid process established soon after birth. In the spring of 1841, I met with an instance in which this lesion was found in a degree so remarkable that it seems difficult to understand how life was continued to the 19th year.

1. On the afternoon of Wednesday the 7th of April 1841, as I was quitting the Hospital about 5 P. M., I found at the inner gate a young man from the country with a face and lips almost purple, labouring under extreme breathlessness, and so enfeebled that he was quite unable to ascend the steps of the staircase. I requested my clerk to get him conveyed to bed; adding, that it was a case of communication of the right and left sides of the heart; and after the patient had a little recovered himself, the following facts were learned.

From infancy this young man, named Andrew Wauchope, had always been subject to shortness of breath, which was very urgent in cold, damp weather. The face, lips, hands, and nails had always been of a purple or livid colour, and though at some times this was less deep than at others, it never entirely disappeared. From his earliest remembrance he was subject to palpitation, or violent knocking action of the heart, which was aggravated to an extreme degree by corporeal exertion, or ascending an acclivity. During the last two years, also, he observed the feet and ankles to be occasionally swelled; but this symptom always subsided. It recurred some time ago, and was considerable at the time of admission.

Notwithstanding these complaints he continued to work at agricultural labour of a light nature, till about a week before the time of admission, when he was attacked with rather copious hemorrhage from the lungs, which took place unexpectedly while he was sitting beside the fire, without any corporeal exertion or mental excitement. The hemorrhage, he added, had recurred several times during the week, though he could not say how frequently, further than that it had sometimes occurred twice daily.

The purple colour of the face, lips, and nails continued; the tongue was of a livid purple colour at the tip, was elsewhere of a leaden tint, except in the centre, where its colour was obscured by a grayish fur of some thickness; the inside of the lips and cheeks were of an unusually deep-blue colour; and the vessels of

the conjunctiva were of a dark blue ; the œdema of the feet and ankles continued.

The anterior part of the chest was very prominent, the sternum arched,* and the shoulders much elevated. Respiration was twenty-eight in the minute performed by the accessory muscles, and with much labour. The sound of respiration was bronchial, blowing and limited, in some parts approaching to cavernous, with some dry sonorous and sibilous wheezing, but no moist rattles.

The cardiac region emitted, on percussion, a preternaturally dull sound ; but above the third rib, on the left side, and on the right side of the sternum, the sound emitted was clear, and in some parts tympanic. The cardiac beats were from 92 to 96 in the minute, performed with rather strong impulse, between the fourth and seventh ribs. The cardiac beats were heard also in the epigastric region, and on the right side of the sternum ; and the first beat was performed with a rough, rasping, or sawing murmur, considerably prolonged.

At this time the diagnosis given was, communication between the right and left chambers of the heart, probably by the *foramen ovale*, hypertrophy of the right ventricle, and probable contraction of the pulmonary artery.

The only treatment consisted in the administration of some laxative medicine, in the observance of perfect rest, and the subsequent limited use of anodynes and antispasmodics.

In the course of four days the breathlessness was greatly abated, and the cough was less frequent and urgent ; the colour of the face, lips, and hands was rather less deeply violet than at admission ; but the vessels of the conjunctiva were still dark-coloured, and the nails had a deep blue tint. The œdema of the feet was diminished ; the urine was increased ; and the pulse was reduced to about 88, but still rose at intervals to 92 and 96. There was no return of hemorrhage.

At this time, when the beats of the heart were again examined, the pulsations were still heard most distinctly on the right side of the sternum ; the first beat was still performed with the rough, rasping murmur, which was also heard most distinctly and forcibly on the right side of the sternum, immediately below the third and fourth rib, and obscuring there the sounds of respiration. The cardiac beats were also heard in the back, but much less forcibly.

From this time he continued much in the same state ; except that when he attempted to rise and move about, the breathlessness was aggravated, and the cheeks and lips became unusually livid.

On the 10th of May, about four weeks after admission, he was attacked in the evening with rigors, followed by discharge of blood from the lungs. This soon stopped, and next day the expectoration only was streaked with blood. The impulse at the heart was strong and labouring, beats 96 in the minute, with the same rasp-

ing murmur, heard on the right side of the sternum, and with a very small pulse at the wrist. The face and lips were again livid; the surface was cold; and the patient was so feeble that it was impossible to examine the chest fully.

On the 14th the coldness of the surface was complained of by the patient himself, the face and extremities were livid, and the breathing became hurried, and progressively increased till the morning of the 15th May, when death took place.

Inspection of the body on the 17th May disclosed the following appearances in the internal organs.

Slight old adhesions were found connecting both pleuræ to the pulmonic pleuræ, mostly cellular, but increasing in firmness towards the convex and posterior part of the chest.

Two ounces of transparent straw-coloured serum were contained in the cavity of the pericardium.

The heart presented no traces of inflammation on its surface or in its substance, but was very much enlarged, especially in its right ventricle and auricle.

The right auricle was a good deal larger than usual; and the membrane forming the *fenestra ovalis* was thin and reticulated, presenting many minute holes; and at its lower *limbus*, close to the tricuspid valve, was a circular or rather elliptical hole, of the diameter of one-third of an inch, communicating directly with the left auricle.

The right ventricle, when cut open, was large in its capacity, and its walls were very much thickened, and considerably firmer than natural. At the apex of the ventricle was found a deposit partly of fibrinous blood, partly of decolorized fibrine, between the walls of the heart and the *columnæ carneæ*. The latter, viz. the fleshy pillars, were much larger and firmer than they usually are in the right ventricle.

The tricuspid valve was rather whiter and more opaque on its ventricular side than usual, but the aperture formed by it was not contracted.

The pulmonary artery was in its diameter much smaller than usual at its origin and connection with the basis of the right ventricle. Its origin was provided not with three distinct semilunar valves, as usual, but with a firm opaque membrane, concave towards the ventricle, convex towards the artery, so elevated into the interior of the artery as to form a truncated cone, with a small aperture in its centre and apex, not larger than to admit a common probe. This aperture was furnished round its margin with an elevated fringe, similar to the warty prominences usually found in diseased valves in general.

Upon examining the convex or arterial side of this membrane, three small membranous slips were observed passing between the membrane and the walls of the artery,—indicating the position of

the attachment of each valve, and at the same time showing that all of them were firmly united to each other. The sinus between two of these slips was deeper than that between the other two.

The weight of the heart was 18 ounces.

The left ventricle and auricle were of the natural size and capacity, and were otherwise healthy.

The apex of the left lung was firm and solid, and contained a cavity the size of a filbert. Several portions at the upper part of the lower lobe were solid, firm, and unusually dark-coloured. Some other small cavities were observed; but upon examining them carefully, they turned out to be dilated parts of the bronchial tubes.

The right kidney was very large, and weighed 9 ounces. Its different textures were pretty natural, excepting at one part, where corresponding to one of the tubular cones at its base, was a dark-coloured patch, in which the striated structure was much darker than usual, and rather obliterated. At another part opposite the base of one of the tubular cones, a similar change had taken place in the striated texture, the latter being much gone, and in its place was substituted a mottled red-coloured granular material.

The left kidney was shrunk and withered, so much as not to weigh more than one ounce and a-half, including the supra-renal capsule. All trace of its original cortical and tubular structure was entirely gone; and all that was left was the calyceine membrane with one or two of the original infundibular holes.

The preparation is preserved in the College Museum.

Some time after, a case with similar obstruction at the valvular apparatus of the pulmonary artery, but much less complete, occurred in the practice of another physician to the Royal Infirmary. Here, however, the aperture in the centre of the membranous diaphragm was at least half-an-inch in diameter.

2. Of this case the following detailed account has since been given me by our former prosecutor, Dr John Reid.

Thomas Shearer, aged 44; admitted on the 14th January 1841, under the care of Dr Graham, by whose permission it is here published.

He was a labourer on the Edinburgh and Glasgow Railway, and was sent from the country as labouring under fever, and was consequently first sent to a fever ward. Next day it was apparent that this was not the case, and he was sent to an ordinary ward; but the breathing was so laborious, and the man's strength was so much impaired, that he was unable to undergo minute examination of the chest.

He could not give any very distinct history of the progress of his disease, but he stated that, about six weeks before his admission, after a debauch of ten days' duration, he was seized with severe cough, which was at first dry, and afterwards attended with profuse expectoration; hoarseness, headach, and thirst superven-

ed, and he gradually lost flesh and strength. When admitted, his intellects appeared somewhat confused ; he complained of frontal headach, and denied that he had any other local uneasiness ; was very hoarse, and spoke very indistinctly, apparently from the very hurried state of his respiration ; frequent cough, without any expectoration ; pulse 120, weak ; tongue thickly coated ; face rather livid. Had twelve leeches to the chest and a blister.

After his admission, the respiration continued very frequent ; about 60 in the minute. He was delirious and restless at night, and died on the 17th.

Sectio Cadaveris, 21st.—Chest.—The left lung appeared somewhat contracted in size, and the inner surfaces of the left pleura were very firmly adherent ; a very large tubercular cavity in the apex of left lung, occupying nearly the whole of the upper lobe ; several large bronchial tubes opened into this cavity ; the rest of the left lung was dense, from the deposition of yellow tubercles, some of which had broken down, forming small *vomicæ*.

The right lung was voluminous ; but the posterior and middle parts of the upper lobe, the posterior part of the middle lobe, and the upper and back portions of the lower lobe, were dense from the deposition of yellow tubercles, and from intercurrent pneumonia. Some of these tubercles were also in the course of breaking down.

The left margin of the outer surface of the pericardium adhered firmly to the costal pleura of the left side.

The heart was increased in size, weighing 13 ounces 2 drachms.

On opening the pericardium, the pulmonary artery and aorta were seen to be much distended, and they gave the sensation to the finger of being full of air. On opening them, a quantity of gas escaped, and also a quantity of frothy blood, with dark-coloured clots floating in it. [This is doubtful to Dr Graham.]

The outer surface of the right auricle, especially the appendix, was covered with a thin layer of firm reticulated lymph, evidently the result of some former inflammation, probably of ancient date. The right auricle was much dilated, and, at the same time, decidedly thicker and firmer than usual. The right ventricle was much hypertrophied, being nearly equal to the usual thickness of the left ventricle, and its cavity was, at the same time, smaller. The tricuspid valve was thickened at some parts, and presented some slight elevations on the inner surface of its lips.

The pulmonary semilunar valves were much thickened, and were so united at their adjacent edges as to form a ring, which could only admit the point of the little finger. This appearance of a ring was only seen on looking downwards from the pulmonary artery, and probably would remain nearly of the same size both during the systole and diastole of the ventricle. The pulmonary artery was much dilated, and its coats attenuated.

The left cavities appeared healthy.

The aortic semilunar valves were somewhat thickened, but were

adequate. The aorta was also dilated at its arch, but not to the same extent as the pulmonary artery; and a number of small yellowish elevations were scattered over its inner surface.

Some small elevations of a pale yellow colour were observed in the site of the elliptical patches at the lower part of the ileum; other abdominal viscera healthy. No gaseous fluid found in any other organ where it does not naturally exist, except in the heart. No appearance of incipient putrefaction anywhere.

Instances, in all respects similar to the lesion now described, and proceeding to the same degree, are rare. Those described by Gintrac are either examples of the narrow formation of the pulmonary artery, or of its termination in a *cul de sac*.

Of the cases most nearly approaching to it one is recorded by Tacconi, in the Bologna Transactions, and another is delineated by Cruveilhier in the 28th number of his Pathological Anatomy, and very briefly described by him.

The case by Tacconi is to the following effect:—

3. A young woman, about the age of 15, was in the practice of moving so slowly that she was obliged to stop every third step. The hands, nails, face, lips, and white of the eyes were remarkably purple. During winter, and when the wind was in the north, she always felt herself upon the point of death. In this state she continued three years from the time when she was first seen by Tacconi, and at length the debility increasing to an extreme degree, and the voice failing, she expired in her 18th year.

Upon inspecting the body the following day, the lungs were found shrunk, with little fluid, and of a very dark colour. About three ounces of viscid dark-coloured blood were scattered over the diaphragm. The heart was of a cubical shape. The left ventricle had the shape of the right; and the right that of the left, being furnished with strong muscular columns. The pulmonary artery was so closed by the mutual adhesion of its three sigmoid valves, that water injected flowed through it only by a very small hole made by a probe previously introduced. The *canalis arteriosus* was closed. But the *foramen ovale*, especially on the side of the right ventricle, was much larger than it is in the fœtus.*

4. In the case given by Cruveilhier, [I use his own words to obviate mis-statement,] “the orifice of the pulmonary artery is singularly narrowed by a species of diaphragm perforated in its centre by a regularly circular aperture about the size of a lentile. This diaphragm, convex at the arterial or upper surface, and concave at the lower or ventricular surface, presents in the former direction three bridges or slips separated by the same number of hollows or sinuses.

“This diaphragm represents the sigmoid valves, and is evidently formed at the expense of these valves.”

* De Bononiensi scientiarum atque artium Instituto atque Academia, Tom. vi. p. 64.

The right ventricle was in this case also hypertrophied, with hypertrophy of the fleshy pillars. But no mention is made of the state of the *foramen ovale* or the *septum cordis*, excepting that the latter was less hypertrophied than the rest of the ventricle. The former was evidently closed.

The principal point of interest in this case is the date of the origin of this peculiar lesion. Was it a congenital malformation, or did it take place some time after birth?

This question it is almost impossible to determine from one case; but various circumstances in the case of Wauchope, and especially the patent condition of the *foramen ovale*, are in favour of its congenital origin, or at least its appearance soon after birth. Cruveilhier allows that the first idea which the appearance of the diaphragm suggests is, that the structure is congenital; and he admits that it is the idea which all the analogous facts collected by other observers have suggested. He adds that it is impossible to admit a congenital lesion of this kind in individuals who have presented symptoms only a short time before death; but allows that where the *foramen ovale* is open, that forms a strong ground for believing that the lesion was congenital.

These considerations, it must be remarked, are all in favour of the inference that this lesion was congenital, or at least originated very soon after birth. The blueness of the face and lips had been observed from the very first; the violent action of the heart had also been an early symptom; and the patient had been subject to short and difficult breathing from infancy. An aperture of considerable size was found establishing a direct and free communication between the right and left auricle; and the membranous diaphragm itself was so firm and complete, that it appeared scarcely possible to believe that it could have been of recent formation.

I have only one other observation to offer on this and similar cases. It has been usually supposed that the open state of the *foramen ovale*, whether direct or oblique, is a primary lesion of the heart, and is pernicious to the patient, in allowing the free intermixture of the blood of the right chambers of the heart with that of the left. Within certain limits this idea is well-founded; and in a certain number of cases, the open state of the *foramen ovale* tends to impede nutrition and to abridge the duration of life. I am, nevertheless, satisfied, both from the facts of the case now related, and several others, of three circumstances; *first*, that the open state of the *foramen ovale* is rarely a primary and solitary lesion; *secondly*, that when it is a solitary lesion it is not injurious, and the venous blood of the right auricle is not thereby necessarily mixed with the arterial blood of the left auricle; and *thirdly*, that, in opposition to what has been hitherto usually taught, the open state of the *foramen ovale* is in a large proportion of cases the means of prolonging life.

It is unnecessary for me to enter into any formal defence of the

latter conclusion, which, however paradoxical it may seem, and however opposed to the usually received dogmas, flows almost directly from the facts which may be traced in every case of open *foramen ovale*. That, in short, is not the primary lesion. From the phenomena of the cases recorded, on the contrary, and from the frequency of the arctated or contracted state of the pulmonary artery, it must be inferred that the obstructed state of that artery is the primary lesion, and determines not only the open state of the *foramen ovale*, but the hypertrophy of the right ventricle. This is the result, whether the pulmonary artery is only greatly narrowed in calibre, or terminates in a *cul de sac*, or is obstructed by a membranous partition formed by the coalition of the semilunar valves.

The effect of such an impediment is manifest. The blood cannot pass into the pulmonary artery with the requisite freedom and facility. The result is over-distension, *first* of the right ventricle, and excessive labour of its muscular apparatus; *secondly*, of the right auricle, and excessive labour of its muscular apparatus, with extreme dilatation of its membranous portion; *thirdly*, over-distension and congestion of the whole venous system all over the body. The lungs, meanwhile, receive little or no blood, and, consequently, the blood is not duly aerated or supplied with oxygen, and cleared of carbon and carbonic acid. This is doubtless an evil and a great one. But Bichat has obscurely suggested, and Dr Williams and Dr Kay have clearly shown, that dark-coloured blood, or that which is venous is adequate to maintain vital action. It is, indeed, a less evil, and more tolerable than total obstruction in any of the large vessels, and especially in a vessel like the pulmonary artery. Every thing that we now know of these cases shows that the obstruction to the circulation through the pulmonary artery must be the main cause of the short and transitory existence of persons labouring under this severe lesion; and that the open state of the *foramen ovale*, instead of being, as William Hunter and other authors imagined, a cause of death, furnishes the only means by which life can be prolonged, while a function so important as that of the circulation through the lungs is impeded.

I am further entitled to infer from various facts in the history of the development of the ovum, that the obstructed, or, it may be, the undeveloped state of the pulmonary artery, is the anatomical cause of the perforated septum, and of the origin of the aorta from the two ventricles when that malformation is observed.

It would lead to some curious and interesting results to inquire by what means the impeded function of the lungs is in these cases compensated; for that it is compensated by the action of the skin and other membranes, can scarcely be doubted. But this would lead me into a field too extensive for consideration at the present time.

